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Cystic Fibrosis in Europe - remote measurement of outcome

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Background

Cystic Fibrosis (CF) occurs randomly in children across all European social strata providing an opportunity to sample their underpinning health care provision for this rare disease in an unbiased manner. We developed a 35 country European cystic fibrosis (CF) demographic registry to compare CF outcomes through an FP6 programme of the EU called EuroCareCF (http://www.eurocarecf.eu).

Methods

We applied methods (http://www.cystic-fibrosis.org.uk) previously used to create country-specific registries after inviting participation through the European CF Society and CF patient organisations using a double hub (Dundee and Prague) and spoke model supplemented by conferences, workshops and telephone support using a single data collection system. Implementing this common data collection platform, we collated demographic and genotype data in around 30,000 patients scattered from Iceland to the Black Sea.

Results

Amongst the ~30,000 CF patients in our Registry, a widely different country-specific prevalence of childhood CF exists that cannot be explained by differences in population size, underlying heterozygote CF gene frequency or under-ascertainment. In particular, we do not believe that the lattermost can explain our findings because in late childhood, we observe a significant paucity of the clinically severe homozygous F508-del form of CF that is of early childhood onset in 90% of cases and is widely dispersed across mainland Europe.

Conclusions

It is likely that an excess premature CF mortality in childhood still occurs across many parts of Europe, a mortality that has largely disappeared in countries such as the UK, France Germany and other wealthy nations. We suggest that much of (better resourced) Western Europe now has a vanishingly low mortality for the severe commonly occurring F508del homozygous CF in childhood that is not replicated in our study in many European countries. The reasons require investigation.

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